

Ascher's syndrome: A rare entity with subclinical hypothyroidism

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Abstract Ascher's or Laffer-Ascher syndrome is characterized by a triad of double upper lip, blepharochalasis, and a nontoxic thyroid enlargement. Here, we present a case of Ascher's syndrome in a 17-year-old male presenting with swelling of lip, blepharochalasis and subclinical hypothyroidism in contrast to nontoxic thyroid enlargement.

Key words

Ascher's syndrome, thyroid, bhepharochalasis.

Introduction

Ascher's or Laffer-Ascher syndrome was first described in 1920 by Ascher.¹ The disease can be inherited but majority are idiopathic in nature.² It characteristically presents as a triad of double upper lip, blepharochalasis, and a nontoxic thyroid enlargement is present in this disorder.³ Many conditions can simulate this disorder including hereditary angioedema, early dermatochalasis, acquired cutis laxa and variants of granulomatous cheilitis.⁴ There are no published case reports of Ascher's syndrome from Nepal till date.

Case Report

A 17-year-old male was referred to me for persistent swelling of eye folds and lips. The swelling was asymptomatic and present for several years. Notably, the swelling was more prominent during early hours of the day. The swelling was not painful but was associated with mild discomfort. Multiple treatments were used

in the past without any relief. No history of any medication use, joint pains or Raynaud's syndrome.

Past history was relatively insignificant. There was no history of such eruptions in other family members. There was no known allergies or atopy in the patient or other families.

On examination, bilateral non-tender soft swelling of periorbital region (**Figure 1**) along with swelling of both lips (**Figure 2**) were noted. Neither increase in local temperature nor any local color changes was noted.

No abnormalities were noted in systemic examination. Complete blood count was within normal limits. There was a rise in ESR of 35mm/1st hour and thyroid stimulating hormone of 5.11 (Normal 0.3-4.5).

Discussion

Ascher's or Laffer-Ascher syndrome was first described in 1920 by Ascher.¹ Only a few hundred cases of this benign condition are reported in the literature. Majority of cases are idiopathic in nature, but it may be inherited as an autosomal dominant disorder.²

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Figure 1 Swelling of bilateral periorbital region.

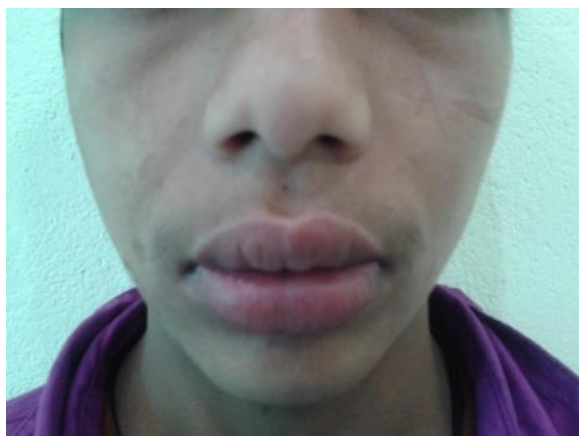


Figure 2 Persistent swelling of both lips.

A triad of double upper lip, blepharochalasis, and a nontoxic thyroid enlargement is present in this disorder. But only 10-50% of patients with Ascher's syndrome have thyroid involvement, and it may not be essential for the diagnosis of Ascher's syndrome.³ In this particular case, there was presence of subclinical hypothyroidism rather than euthyroid goiter as mentioned in the literature.

Blepharochalasis is described as a laxity of the skin of the upper eyelid, causing the tissue between the eyebrows and the lid edge to hang loosely over the lid margins.⁴ In more than 80% of cases swelling of lip and eyelid occur at about

the same time and usually manifest before the age of 20.⁴

The histologic findings are prominent minor salivary glands and a mixed inflammatory cell infiltrate.⁵

The differential diagnosis includes hereditary angioedema, early dermatochalasis, acquired cutis laxa and variants of granulomatous cheilitis. So, keeping this disease in mind would prevent making it several misdiagnosis.⁴ It is imperative to follow up these patients, so timely surgery can be done.⁶ The treatment of choice is generally cosmetic surgery.⁷

References

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